SLICC/ACR Damage Index is valid, and renal and pulmonary organ scores are predictors of severe outcome in patients with systemic lupus erythematosus

Stoll, T ; Seifert, Burkhardt ; Isenberg, D A

Abstract: We investigated the Systemic Lupus International Collaborative Clinics/American College of Rheumatology (SLICC/ACR) Damage Index as a predictor of severe outcome and an indicator of morbidity in different ethnic groups, and in regard to its validity. We retrospectively studied disease course within 10 yr of diagnosis in an inception cohort of 80 patients with systemic lupus erythematosus (SLE). The mean renal damage score (DS) at 1 yr after diagnosis was a significant predictor of endstage renal failure and the mean pulmonary DS at 1 yr significantly predicted death within 10 yr of diagnosis. Compared to Caucasians, Afro-Caribbeans and Asians had significantly higher mean total DS at 5 and 10 yr, and higher mean renal DS at 10 yr. At 5 yr, the mean renal DS in Afro-Caribbeans and the mean neuropsychiatric DS in Asians were significantly higher than in Caucasians. The rate of endstage renal failure in Caucasians was significantly lower than in the other ethnic groups. Our results confirm the validity of the SLICC/ACR Damage Index.

DOI: https://doi.org/10.1093/rheumatology/35.3.248

British Journal of Rheumatology 1996;35:248–254

SLICC/ACR DAMAGE INDEX IS VALID, AND RENAL AND PULMONARY ORGAN SCORES ARE PREDICTORS OF SEVERE OUTCOME IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

T. STOLL, B. SEIFERT* and D. A. ISENBERG

*Bloomsbury Rheumatology Unit/Division of Rheumatology, Department of Medicine, University College of London, London W1P 9PG and *Department of Biostatistics, ISPM, University, CH-8091 Zürich, Switzerland*

SUMMARY

We investigated the Systemic Lupus International Collaborative Clinics/American College of Rheumatology (SLICC/ACR) Damage Index as a predictor of severe outcome and an indicator of morbidity in different ethnic groups, and in regard to its validity. We retrospectively studied disease course within 10 yr of diagnosis in an inception cohort of 80 patients with systemic lupus erythematosus (SLE). The mean renal damage score (DS) at 1 yr after diagnosis was a significant predictor of endstage renal failure and the mean pulmonary DS at 1 yr significantly predicted death within 10 yr of diagnosis. Compared to Caucasians, Afro-Caribbeans and Asians had significantly higher mean total DS at 5 and 10 yr, and higher mean renal DS at 10 yr. At 5 yr, the mean renal DS in Afro-Caribbeans and the mean neuropsychiatric DS in Asians were significantly higher than in Caucasians. The rate of endstage renal failure in Caucasians was significantly lower than in the other ethnic groups. Our results confirm the validity of the SLICC/ACR Damage Index.

KEY WORDS: SLICC/ACR Damage Index, Death, Endstage renal failure, Morbidity in different ethnic groups, SLE.

THE Systemic Lupus International Collaborative Clinics/American College of Rheumatology (SLICC/ACR) Damage Index, developed in 1992, was created to assess persistent reflection of disease activity in SLE patients and has been used in various 'paper' and real patient studies [1]. Damage, that means irreversible impairment, was defined as being continuously persistent for at least 6 months. The index is shown in Table Ia and b. In 1994, a weighted scoring system for SLICC/ACR Damage Index was presented [2], based upon the opinion of lupus experts. Our retrospective study, based on the SLICC/ACR Damage Index, was undertaken to investigate the following four questions. Firstly can minor damage predict progression to major damage? For example, parts of the renal damage score (DS), such as a reduced glomerular filtration rate and proteinuria, have been shown to be associated with the subsequent development of renal insufficiency [3, 4]. Similarly, prediction of death by the SLICC/ACR Damage Index is also of interest. Secondly, can morbidity becompared between different ethnic groups? As survival in SLE patients has improved, comparison of mortality between ethnic groups has become less sensitive, but an analysis of morbidity may reveal other differences. Thus, the SLICC/ACR Damage Index, as an instrument of outcome assessment during lifetime, seems likely to satisfy this purpose. Higher mortality rates in Afro-Caribbeans [5, 6] and Asians [7] compared to Caucasians could reflect either more severe disease, higher disease prevalence [8, 9] or both and could be paralleled by higher DS. Thirdly, would the answers to the first two questions be in accordance with the literature and therefore demonstrate the validity of the SLICC/ACR Damage Index? Fourthly, would the weighted total DS [2] provide more insights into systemic lupus erythematosus (SLE) than the unweighted total DS?

Submitted 6 July 1995; revised version accepted 2 October 1995.

Correspondence to: T. Stoll, Department of Rheumatology, City Hospital Waid, Tièchestr. 99, CH 8037 Zürich, Switzerland.

PATIENTS AND METHODS

An inception cohort of all of our SLE patients in whom the diagnosis was established at least 10 yr prior to November 1994, and who had subsequently attended a specialist lupus clinic, was studied. At each out-patient consultation, detailed clinical and serological information was recorded (and kept separate from the hospital notes, although these were also available for perusal). The SLICC/ACR Damage Index was assessed retrospectively from the notes 1, 5 and 10 yr after diagnosis by the same rheumatologist (TS). In the case of a lethal outcome, the last DS was determined 6 months before death. The weighted total DS was calculated by multiplying the neuropsychiatric, renal and cardiovascular DS by four, the ocular, pulmonary, peripheral vascular and malignancy DS by three, and the gastrointestinal and musculoskeletal DS by two, resulting in a potential maximum of 133 points [2]. The date at which a patient fulfilled the fourth SLE classification criterion [10] was recorded as the date of diagnosis. Every patient was carefully reviewed for evidence of having had a severe outcome within 10 yr after diagnosis, defined as either death or endstage renal failure (ESRF) necessitating dialysis. If a patient had moved and had not been attending our out-patient clinic, attempts were made to contact her/him and their physicians to obtain the necessary data.

TABLE Ia

SLICC/ACR Damage Index. Definition of damage: occurring since onset of lupus, ascertained by clinical assessment and present for at least 6 months unless otherwise stated. Repeat episodes mean at least 6 months apart to score 2. The same lesion cannot be scored twice. A precise definition of items is contained in a glossary

Organ (system)	Item	Score [maximal]
Ocular	Any cataract ever	1
	Retinal change OR optic atrophy	1 [2]
Neuropsychiatric	Cognitive impairment OR Major psychosis	1
	Seizures requiring therapy for ≥6 months	1
	Cerebral vascular accident ever (Score 2 if >1×) OR resection not for malignancy	1 (2)
	Cranial or peripheral neuropathy (excluding optic)	1
	Transverse myelitis	1 [6]
Renal	Estimated or measured GFR < 50%	1
	Proteinuria ≥3.5 g/24 h	1
	ESRF (regardless of dialysis or transplantation)*	1–3* [3]
Pulmonary	Pulmonary hypertension (right ventricular prominence or loud P2)	1
	Pulmonary fibrosis (clinically and/or by X-ray)	1
	Shrinking lung (by X-ray)	1
	Pleural fibrosis (by X-ray)	1
	Pulmonary infarction (by X-ray) OR resection not for malignancy	1 [5]
Cardiovascular	Angina OR Coronary artery bypass	1
	Myocardial infarction ever (Score 2 if >1×)	1 (2)
	Cardiomyopathy (ventricular dysfunction)	1
	Valvular disease (diastolic murmur, or systolic murmur > 3/6)	1
	Pericarditis OR Pericardiectomy	1 [6]
Peripheral vascular	Claudication	1
	Minor tissue loss (pulp space)	1
	Significant tissue loss ever (at least loss or resection of a digit) (Score 2 if >1×)	1 (2)
	Venous thrombosis with swelling, ulceration OR venous stasis	1 [5]
Gastrointestinal	Infarction or resection of bowel below duodenum, spleen, liver or gall bladder ever (Score 2 if >1 site)	1 (2)
	Mesenteric insufficiency	1
	Chronic peritonitis	1
	Stricture OR Upper gastrointestinal tract surgery ever	1
	Pancreatic insufficiency requiring enzyme replacement OR with pseudocyst	1 [6]
Musculoskeletal	Muscle atrophy OR weakness	1
	Deforming or erosive arthritis (including reducible deformities, excluding AVN)	1
	Osteoporosis with fracture or vertebral collapse (excluding AVN)	1
	Avascular necrosis = AVN (Score 2 if >1×)	1 (2)
	Osteomyelitis	1 [6]
Skin	Scarring chronic alopecia	1
	Extensive scarring of panniculum other than scalp and pulp space	1
	Skin ulceration (excluding thrombosis) of more than 6 months	1 [2]
Gonadal	Premature gonadal failure	1 [1]
Endocrine	Diabetes requiring therapy, regardless of treatment	1 [1]
Malignancy	Malignancy (exclude dysplasia) (Score 2 if >1 site)	1 (2) [2]

Potential maximum of the total ADI: 46 points.
*Adding up to a total renal DS of 3 points.

TABLE Ib

SLICC/ACR Damage Index: Glossary of terms

Damage:
Non-reversible change, not related to active inflammation, occurring since onset of lupus, ascertained by clinical assessment and present for at least 6 months unless otherwise stated. Repeat episodes mean at least 6 months apart to score 2. The same lesion cannot be scored twice.

Cataract:
A lens opacity in either eye, ever, whether primary or secondary to steroid therapy, documented by ophthalmoscopy.

Retinal change:
Documented by ophthalmoscopic examination, may result in field defect, legal blindness.

Optic atrophy:
Documented by ophthalmoscopic examination.

Cognitive impairment:
Memory deficit, difficulty with calculation, poor concentration, difficulty in spoken or written language, impaired performance level, documented on clinical examination or by formal neurocognitive testing.

Major psychosis:
Altered ability to function in normal activity due to psychiatric reasons. Severe disturbance in the perception of reality characterized by the following features: delusions, hallucinations, incoherence, marked loose associations, impoverished thought content, marked illogical thinking, bizarre, disorganized or catatonic behaviour.

Seizures:
Paroxysmal electrical discharge occurring in the brain and producing characteristic physical changes including tonic and clonic movements and certain behavioural disorders. Only seizures requiring therapy for 6 months are counted as damage.

CVA:
Cerebral vascular accident resulting in focal findings such as paresis, weakness etc., OR surgical resection for causes other than malignancy.

Neuropathy:
Damage to either a cranial or a peripheral nerve (excluding optic), resulting in either motoric or sensory dysfunction.

Transverse myelitis:
Lower extremity weakness or sensory loss with loss of rectal and urinary bladder sphincter control.

Glomerular filtration rate (GFR):
Estimated or measured GFR <50%.

Cardiovascular:
Myocardial infarction (documented by EKG and enzymes), ever. Cardiomyopathy (ventricular dysfunction documented clinically).

Peripheral vascular:
Claudication persistent for 6 months, by history. Minor tissue loss, such as pulp space, ever. Significant tissue loss, such as loss of digit or limb, or resection, ever. Venous thrombosis with swelling, ulceration, or clinical evidence of venous stasis.

continued

continued overleaf

TABLE Ib—*continued*
SLICC/ACR Damage Index: Glossary of terms

Gastrointestinal:
Infarction or resection of bowel below duodenum, by history. Resection of spleen, liver or gall bladder ever, for whatever cause. Mesenteric insufficiency, with diffuse abdominal pain, on clinical examination. Chronic peritonitis, with persistent abdominal pain and peritoneal irritations, on clinical examination. Oesophageal stricture, shown on endoscopy. Upper gastrointestinal tract surgery, such as correction of stricture, ulcer surgery etc., ever, by history.

Musculoskeletal:
Muscle atrophy or weakness, demonstrated on clinical examination. Deforming or erosive arthritis, including reducible deformities, excluding avascular necrosis, on clinical examination. Osteoporosis with fracture or vertebral collapse (excluding avascular necrosis), demonstrated on X-ray. Avascular necrosis, demonstrated on any image technique. Osteomyelitis, documented clinically, and supported by culture evidence.

Skin:
Scarring chronic alopecia, documented clinically. Extensive scarring or panniculum other than scalp and pulp space, documented clinically. Skin ulceration (excluding thrombosis).

Premature gonadal failure:
Secondary amenorrhoea.

Malignancy:
Documented by pathology, excluding dysplasias.

Statistical analysis

The survival times to severe outcome were analysed using Cox regression. Damage scores and severe outcomes, respectively, were compared beween different ethnic groups by the Kruskal–Wallis rank test and Fisher's exact test for contingency tables. In case of an overall difference at $P \leqslant 0.1$, pairwise differences were tested using the Mann–Whitney rank and Fisher's exact test. The analyses were performed using Statview 4.02 and Splus 3.2. Differences were considered significant at $P < 0.05$, and no adjustment was made for multiple outcome testing.

RESULTS

Eighty patients, 77 females and three men, aged 33.2 ± 12 yr (mean $\pm$ S.D.), fulfilling the classification criteria of SLE [10] were studied. Fifty-three were Caucasians, 15 Afro-Caribbeans, nine Asians and three were of mixed ethnic origin. Damage could be scored at 1 yr after diagnosis in all 80 patients and at 5 yr in 76 patients; three had died and one had moved away. At 10 yr, the DS could be determined in 68 patients—six had died and six had moved away. Thus, by the end of the study, 10 yr after establishing the diagnosis of

TABLE II
SLE patients who died

Patient	Age (yr) at diagnosis	Pulmonary DS at 1 yr*	Cause of death (important comorbidity)	Years from diagnosis to death
♀, Afro-Caribbean	31.6	0	Myocardial infarction (ESRF)	4
♀, Afro-Caribbean	15.1	0	Myocarditis	1.2
♀, Afro-Caribbean	14.8	0	Peritoneal sepsis (ESRF)	8.8
♀, Afro-Caribbean	35.2	1†	Thrombotic thrombopenic purpura	0.8
♀, Caucasian	48.8	0	Hepatic failure	9.7
♀, Caucasian	24.3	0	Thrombotic thrombopenic purpura	5
♀, Caucasian	31.4	0	Myocardial infarction	8.1
♀, Caucasian	52.4	0	Sepsis (Hodgkin's lymphoma)	3.4
♀, Caucasian	27.5	0	Massive pulmonary embolism	7
♀, Caucasian	21.6	1‡	Cardiac arrest	0.6

*Pulmonary damage score 1 yr after diagnosis, or in case of a lethal outcome earlier than 1.5 yr after diagnosis 6 months before death, respectively.
†Pulmonary fibrosis.
‡Pulmonary hypertension.

TABLE III
Progression of damage scores and cumulative prevalence of damage* over time (* = percentage of patients with a DS > 0)

Time after diagnosis	DS: mean (range); cumulative prevalance of damage at 1 yr	DS: mean (range); cumulative prevalance of damage at 5 yr	DS: mean (range); cumulative prevalance of damage at 10 yr
Total damage score (unweighted)	0.40 (0–3); 32.5%	0.75 (0–4); 51.3%	1.50 (0–5); 67.6%
Weighted damage score (total)	1.19 (0–8); 32.5%	2.32 (0–13); 51.3%	4.81 (0–15); 67.6%
Ocular damage score	0.00 (0); 0%	0.00 (0); 0%	0.04 (0–1); 3.8%
Neuropsychiatric damage score	0.08 (0–1); 7.5%	0.13 (0–1); 13.2%	0.24 (0–2); 22.1%
Renal damage score	0.05 (0–1); 5%	0.24 (0–3); 17.1%	0.53 (0–3); 32.4%
Pulmonary damage score	0.05 (0–1); 5%	0.04 (0–1); 3.9%	0.09 (0–2); 7.4%
Cardiovascular damage score	0.03 (0–1); 2.5%	0.04 (0–1); 3.9%	0.09 (0–1); 8.8%
Peripheral vascular damage score	0.03 (0–1); 2.5%	0.08 (0–1); 6.6%	0.12 (0–2); 10.3%
Gastrointestinal damage score	0.03 (0–1); 2.5%	0.04 (0–1); 3.9%	0.03 (0–1); 2.9%
Musculoskeletal damage score	0.11 (0–1); 11.3%	0.15 (0–2); 13.2%	0.31 (0–2); 22.1%
Skin damage score	0.04 (0–1); 3.8%	0.07 (0–1); 6.6%	0.06 (0–1); 5.9%
Gonadal damage score	0.00 (0); 0%	0.01 (0–1); 1.3%	0.03 (0–1); 2.9%
Endocrine, malignancy damage score	0.00 (0); 0%	0.00 (0); 0%	0.00 (0); 0%

TABLE IV
Damage scores [only DS of organ (systems) with statistically significant differences shown] in different outcome groups at 1, 5 and 10 yr after diagnosis

	Damage score at											
	1 yr				5 yr				10 yr			
Outcome	Total	Weighted	Renal	Pulmonary	Total	Weighted	Renal	Pulmonary	Total	Weighted	Renal	Pulmonary
Survival, DS												
Mean	0.37	1.14	0.04	0.03‡	0.70	2.19	0.20	0.03	1.52	4.83	0.53	0.06*
Range	0–3	0–8	0–1	0–1	0–3	0–12	0–3	0–1	0–5	0–15	0–3	0–1
Death, DS												
Mean	0.60	1.50	0.10	0.20‡	1.29	3.57	0.57	0.14	1.25	4.50	0.50	0.50*
Range	0–1	0–4	0–1	0–1	0–4	0–13	0–3	0–1	0–2	0–8	0–2	0–2
No ESRF, DS												
Mean	0.39	1.16	0.03†	0.05	0.69*	2.04†	0.14‡	0.04	1.40*	4.30†	0.35†	0.10
Range	0–3	0–8	0–1	0–1	0–3	0–12	0–2	0–1	0–5	0–15	0–2	0–2
ESRF, DS												
Mean	0.50	1.50	0.33†	0.00	1.50*	5.50†	1.33‡	0.00	2.80*	11.20†	2.80†	0.00
Range	0–1	0–4	0–1	0	0–4	0–13	0–3	0	2–3	8–12	2–3	0

*$P < 0.05$; †$P < 0.01$; ‡$P < 0.001$.

SLE, 10 patients had died, on average 4.9 ± 3.4 yr after diagnosis, at a mean age of 35.1 yr (in those four patients who died between 7 and 9.7 yr after diagnosis, a '10 yr DS' 6 months before death was determined). By 10 yr, six patients had developed ESRF, on average 7.2 ± 3 yr after diagnosis, at a mean age of 36.9 yr. Table II shows the data of the 10 patients who died. DS progressed with disease duration (Table III). A decrease of DS, such as in the pulmonary, gastrointestinal or skin item of the SLICC/ACR Damage Index, reflects the finding that some patients with a higher DS had either died or moved away (Table III).

Damage scores in different outcome groups

In 10 and six patients, a search was made for predictors for two major outcomes within the first 10 yr after diagnosis: death and ESRF (Table IV). The mean pulmonary DS at 1 yr in surviving patients was significantly lower compared to those who had died ($P = 0.0095$). The mean renal DS at 1 yr of those patients developing ESRF was significantly higher than the corresponding mean in the patients with no ESRF up to 10 yr after diagnosis ($P = 0.008$). The increased renal DS at 1 yr in those patients developing ESRF was due to proteinuria ⩾3.5 g/day. In those two patients with a renal DS of 0 at 5 yr, at the end of the second year after diagnosis a proteinuria of 0.5 and 1.1 g/day, respectively, as expression of lupus nephritis was present, but proteinuria in this range does not score as damage. The earliest ESRF was observed 2.96 yr after diagnosis. Figures 1 and 2 depict all these findings as Kaplan–Meier survival curves. Table IV also outlines the significant differences in pulmonary, renal, total and weighted DS at 5 and 10 yr beween outcome groups. The strong association of renal DS and ESRF at 5 and at 10 yr was to be expected as ESRF is identical with a renal DS of 3.

Damage scores and outcomes in different ethnic groups

In the comparison between ethnic groups, patients of mixed origin were not included in the analysis because of their small number ($n = 3$). Age at diagnosis did not differ between Caucasians ($n = 53$; age at diagnosis 35 ± 12.7 yr), Afro-Caribbeans ($n = 15$; 30.9 ± 11.5 yr) and Asians ($n = 9$; 28.7 ± 6.5 yr) ($P = 0.27$). No

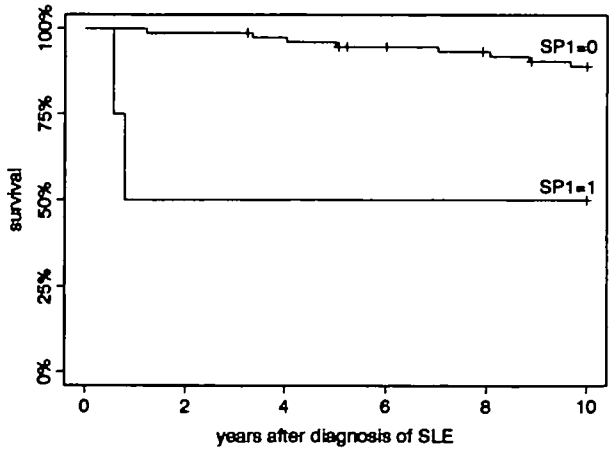

FIG. 1.—Survival (Kaplan–Meier) depending on the pulmonary DS at 1 yr (SP1).

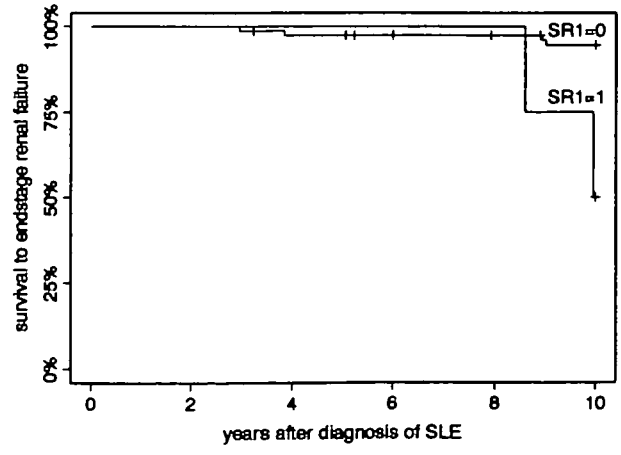

FIG. 2.—Survival to endstage renal failure (Kaplan–Meier) depending on the renal DS at 1 yr (SR1).

statistically significant differences could be observed 1 yr after diagnosis. However, the mean total and renal DS at 5 yr in Afro-Caribbeans were significantly higher than in Caucasians ($P = 0.049$ and $P = 0.036$, respectively). The mean weighted and neuropsychiatric DS at 5 yr in Asians were significantly higher than in Caucasians ($P = 0.034$ and $P = 0.027$, respectively). Thus, the development of significant differences occurred between 1 and 5 yr after diagnosis. The differences between ethnic groups at 10 yr resembled those noticed after 5 yr: mean total, weighted and renal DS in Afro-Caribbeans and Asians were significantly higher than in Caucasians (see Table V). Moreover, Caucasians developed ESRF less frequently than patients of other ethnic origins ($P = 0.006$). This correlates with the finding that at 10 yr renal damage was significantly less common in Caucasians (20.8%) than in Afro-Caribbeans (63.6%) and Asians (57.1%) ($P = 0.006$).

Comparison of total and weighted damage score

Table IV demonstrates that there was slightly more statistical power resulting from the comparison of weighted DS than from the comparison of total DS: $P = 0.008$ and $P = 0.007$ *vs* $P = 0.034$ and $P = 0.034$ (outcome ESRF, 5 and 10 yr after diagnosis). The sole statistically significant difference between Caucasians and Afro-Caribbeans at 5 yr was observed when total DS were compared, whereas between Caucasians and Asians at 5 yr only weighted DS differed significantly.

DISCUSSION

By definition, the SLICC/ACR Damage Index scores cumulative damage since the onset of SLE and is therefore likely to increase with disease progression [1].

The mean pulmonary DS 1 yr after diagnosis significantly predicted death. The relevant items in those who died were either pulmonary fibrosis or pulmonary hypertension (Table II). This was predictable, as interstitial lung disease [11] and pulmonary hypertension in SLE [12] are known to increase mortality. The mean renal DS at 1 yr significantly correlated with the development of ESRF. This corresponds with the known prognostic value of reduced glomerular filtration rate or proteinuria for poor renal outcome [3, 4]. Moreover, significantly higher renal DS after 5 and 10 yr in patients with ESRF appear to corroborate these findings. It is noteworthy that the significant differences in mean total and weighted DS at 5 and 10 yr seem to reflect to a great extent the differences in renal DS. These two findings, being in accordance with the literature [3, 4, 11, 12], demonstrate prognostic validity for the pulmonary and renal item of the SLICC/ACR Damage Index.

Other organ DS, e.g. the neuropsychiatric or the cardiovascular, or the total DS did not show prognostic value in the present study.

Our report is the first to describe ethnic differences by means of the SLICC/ACR Damage Index (Table V). Significant differences in renal, neuropsychiatric and total DS developed between 1 and 5 yr after diagnosis, as have been described previously for both renal [13] and neuropsychiatric involvement [14, 15] within 5 yr of diagnosis. However, our data suggest that disease progression within the first 10 yr in Caucasians is slower than for Afro-Caribbeans or Asians. Compared to Caucasians, significantly higher mean renal DS at 5 and 10 yr in Afro-Caribbeans [6, 16, 17], and at 10 yr in Asians [18, 19], respectively, generally correspond to the literature. The significantly lower occurrence of ESRF in Caucasians, compared to the other ethnic groups, confirms these results. Moreover, a higher mean neuropsychiatric DS 5 yr after diagnosis in Asians than in Caucasians was found. This difference, which is no longer significant at 10 yr, may have occurred by chance. These data demonstrate the discriminatory validity of the renal item of the SLICC/ACR Damage Index, as most significant differences between ethnic groups are corroborated by being present on at least two assessments (Tables IV and V) and above all are in accordance with reports from other groups [13–19].

Statistical power was slightly increased when using the weighted instead of the total DS in terms of comparing different outcomes overall, though not for comparison of ethnic groups (Tables IV and V). Therefore, the weighted DS offers little advantage compared to the total DS.

We noted a rather low prevalence of different organ involvement, e.g. 13% neuropsychiatric disease at 5 yr and 24% at 10 yr after diagnosis compared to figures of CNS involvement of 12–59% reported by other groups [20]. However, the SLICC/ACR Damage Index scores only those items likely to be relevant to outcome. Migraine, a frequent neuropsychiatric finding in SLE patients, will not score as damage. Moreover, a symptom/finding has to be present continuously for at least 6 months to score. Finally, assessment by the SLICC/ACR Damage Index is designed to be feasible for all physicians to complete, being based on clinical examination and simple investigations (urinalysis, creatinine, X-ray of the chest). Subtle changes such as cognitive impairment will only be scored when they are clinically overt. However, such impairment demonstrable with detailed neuropsychiatric testing or MRI will not be counted as damage in a patient who seems clinically unaffected.

In conclusion, in this study a severe outcome in patients with SLE correlates significantly with a higher mean renal or pulmonary DS at 1 yr. Renal (and total) DS at 5 and 10 yr after diagnosis were significantly higher in Asians and Afro-Caribbeans than in Caucasians. Moreover, as our results are in accordance with the literature prior to the use of the SLICC/ACR Damage Index, they confirm the validity of the SLICC/ACR Damage Index, especially of its renal and pulmonary items. However, because of the size of the studied retrospective cohort, these findings warrant further evaluation in prospective studies with greater numbers of patients.

TABLE V

Total, weighted, renal neuropsychiatric (= neurop.) and pulmonary (= pulm.) damage in different ethnic groups at 1, 5 and 10 yr [only DS of organ (systems) with statistically significant differences and pulmonary DS shown]

	Damage score at														
	1 yr					5 yr					10 yr				
Ethnic group	Total	Weighted	Renal	Neurop.	Pulm.	Total	Weighted	Renal	Neurop.	Pulm.	Total	Weighted	Renal	Neurop.	Pulm.
Caucasians, DS															
Mean	0.32	1.02	0.04	0.06	0.04	0.56*	1.69*	0.12*	0.08*	0.04	1.15‡†	3.60‡†	0.27†*	0.17	0.1
Range	0–2	0–8	0–1	0–1	0–1	0–2	0–8	0–1	0–1	0–1	0–5	0–15	0–2	0–1	0–2
Afro-Caribbeans, DS															
Mean	0.67	1.73	0.07	0.13	0.07	1.42*	4.58	0.75*	0.17	0	2.55 ‡	8.73 ‡	1.36†	0.36	0
Range	0–3	0–7	0–1	0–1	0–1	0–4	0–13	0–3	0–1	0	2–3	5–12	0–3	0–1	0
Asians, DS															
Mean	0.56	1.67	0.11	0.11	0.11	1.11	3.56*	0.22	0.33*	0.11	2.57†	8.14†	1.00*	0.43	0.14
Range	0–2	0–5	0–1	0–1	0–1	0–3	0–8	0–1	0–1	0–1	1–4	2–12	0–3	0–2	0–1
Mixed,§ DS															
Mean	0	0	0	0	0	0.33	0.33	0.33	0.33	0	0.50	0.50	0.50	0.50	0
Range	0	0	0	0	0	0–1	0–1	0–1	0–1	0	0–1	0–1	0–1	0–1	0

*$P < 0.05$; †$P < 0.01$; ‡$P < 0.001$.

§Because of the small number ($n = 3$), not included in statistical analysis.

Acknowledgements

We wish to thank for their support: Ciba-Geigy-Jubiläums-Stiftung, CH-4002 Basel, Theodor und Ida Herzog-Egli-Stiftung, CH-8008 Zürich, and Foundation for specialists training of the Swiss Society of Rheumatology and Physical Medicine, CH-8038 Zürich, Switzerland.

References

1. Gladman D, Ginzler E, Goldsmith C *et al.* Systemic lupus international collaborative clinics: development of a damage index in systemic lupus erythematosus. *J Rheumatol* 1992;**19**:1820–1.
2. Gladman D, Goldsmith C, Urowitz M *et al.* Derivation of a weighting scale for the SLICC/ACR damage index—preliminary results. *Arthritis Rheum* 1994;**37**(suppl. 9): S326 (Abstract 994).
3. Levey AS, Lan SP, Corwin HL *et al.* Progression and remission of renal disease in the lupus nephritis collaborative study. *Ann Intern Med* 1992;**116**:114–23.
4. Esdaile JM, Abrahamowicz M, MacKenzie T, Hayslett JP, Kashgarian M. The time-dependence of long-term prediction in lupus nephritis. *Arthritis Rheum* 1994; **37**:359–68.
5. Siegel M, Lee SL. The epidemiology of systemic lupus erythematosus. *Semin Arthritis Rheum* 1973;**3**:1–54.
6. Reveille JD, Bartolucci A, Alarcón GS. Prognosis in SLE. Negative impact of increasing age at onset, black race and thrombocytopenia, as well as causes of death. *Arthritis Rheum* 1990;**33**:37–48.
7. Kaslow RA. High rate of death caused by systemic lupus erythematosus among U.S. residents of Asian descent. *Arthritis Rheum* 1982;**25**:414–8.
8. Johnson AE, Gordon C, Palmer RG, Bacon PA. The prevalence and incidence of systemic lupus erythematosus in Birmingham, England. *Arthritis Rheum* 1995;**38**: 551–8.
9. Serdula MK, Rhoads GG. Frequency of systemic lupus erythematosus in different ethnic groups in Hawaii. *Arthritis Rheum* 1979;**22**:328–33.
10. Tan EM, Cohen AS, Fries JF *et al.* The 1982 revised criteria for the classification of systemic lupus erythematosus. *Arthritis Rheum* 1982;**25**:1271–7.
11. Weinrib L, Sharma OP, Quismorio FP Jr. A long term study of interstitial lung disease in systemic lupus erythematosus. *Semin Arthritis Rheum* 1990;**20**:48–56.
12. Asherson RA, Higenbottam TW, Dinh Xuan AT, Khamashta MA, Hughes GRV. Pulmonary hypertension in a lupus clinic: experience with twenty-four patients. *J Rheumatol* 1990;**17**:1292–8.
13. Adelman DC, Wallace DJ, Klinenberg JR. Thirty-four-year delayed-onset lupus nephritis: a case report. *Arthritis Rheum* 1987;**30**:479–80 (letter).
14. Feinglass EJ, Arnett FC, Dorsch CA, Zizic TM, Stevens MB. Neuropsychiatric manifestations of systemic lupus erythematosus: diagnosis, clinical spectrum and relationship to other features of the disease. *Medicine* 1976; **55**:323–39.
15. Seibold JR, Medsger TA Jr, Buckingham RB, Kelly RH. Central nervous system (CNS) involvement in systemic lupus erythematosus (SLE). Clinical features and response to therapy. *Arthritis Rheum* 1981;**24**(suppl. 4): S70 (Abstract 76).
16. Ward MM, Studenski S. Clinical manifestations of systemic lupus erythematosus. Identification of racial and socioeconomic influences. *Arch Intern Med* 1990;**150**: 849–53.
17. Hochberg MC, Boyd RE, Ahearn JM *et al.* Systemic lupus erythematosus: a review of clinico-laboratory features and immunogenetic markers in 150 patients with emphasis on demographic subsets. *Medicine* 1985;**64**: 285–95.
18. Lee HS, Spargo BH. A renal biopsy study of lupus nephropathy in the United States and Korea. *Am J Kidney Dis* 1985;**5**:242–50.
19. Pistiner M, Wallace DJ, Nessim S, Metzger AL, Klinenberg JR. Lupus erythematosus in the 1980s: A survey of 570 patients. *Semin Arthritis Rheum* 1991;**21**: 55–64.
20. Wallace DJ. The clinical presentation of SLE. In: Wallace DJ, Hahn BH, eds. *Dubois' lupus erythematosus*, 4th edn. Philadelphia and London: Lea and Febiger, 1993:317–21.